CLINICO-PATHOLOGICAL STUDY OF CONGENITAL CYSTIC ADENOMATOID MALFORMATION

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CERTIFICATE

This is to certify that the dissertation entitled **"CLINICO-PATHOLOGICAL STUDY OF CONGENITAL CYSTIC ADENOMATOID MALFORMATION"** is a bonafide work done by **Dr.Venkatasaravanan .S** under my guidance and supervision during the period between 2007 – 2010 towards the partial fulfillment of requirement for the award of M.Ch Branch V (Paediatric Surgery) degree examination held in August 2010 by The Tamilnadu Dr. M.G.R. Medical University, Chennai.

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DECLARATION

I solemnly declare that the dissertation entitled **"CLINICO-PATHOLOGICAL STUDY OF CONGENITAL CYSTIC ADENOMATOID MALFORMATION"** is the original work done by me at The Institute of Child Health & Hospital for Children, Egmore, during the M.Ch. course (2007 to 2010), under the guidance and supervision of Prof. S.V. Senthilnathan MS., M.Ch., Professor and H.O.D. of Paediatric Surgery. The dissertation is submitted to THE TAMILNADU Dr. M.G.R. MEDICAL UNIVERSITY towards the partial fulfillment of requirement for the award of M.Ch. (BRANCH – V) IN PAEDIATRIC SURGERY.

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CONTENTS

INTRODUCTION

Congenital cystic adenomatoid malformation (CCAM) is a rare developmental abnormality of the lung.They are similar to benign lung tumors (1).The underlying feature of a Congenital cystic adenomatoid malformation is an excessive over growth of terminal respiratory bronchioles, forming cysts of various sizes (1).This abnormal lung tissue is of defective epithelial-mesenchymal architecture(1,57)

The congenital lesions are detectable on prenatal ultrasound .They appear as solid or cystic masses.Mode of presentation of CCAM vary widely in antenatal and neonatal period.

AIM OF THE STUDY

- To analyze the effectiveness of ante-natal ultrasonogram
- To study the age of presentation of congenital cystic adenomatoid malformation
- To analyze the various modes of presentation of congenital cystic adenomatoid malformation
- To analyze the pathology of these lesions
- Follow up of the cases during the study period

REVIEW OF LITERATURE

Congenital lung abnormalities are uncommon and diverse in their presentation. However, all those who care for infants and children must have an appreciation for the diagnosis and treatment of these abnormalities because the potential consequencies can be life threatening. Inorder to understand the pathophysiology of these malformations ,a basic understanding of lung development as well as respiratory physiology and anatomy is required and these will be discussed in this review of literature.

EMBRYOLOGY OF LUNG DEVELOPMENT

During the third week of gestation ,the human embryo develops a diverticulum of the ventral foregut ,which forms the primordium of the respiratory system.This is mainly endodermal in origin;however ,cartilaginous and muscular elements will be derived from the splanchnic mesoderm that surrounds the primitive foregut.As the respiratory diverticulum grows caudad ,it becomes separated from the foregut by the lateral esophageal ridges ,which fuse to form a septum at the end of fourth gestational week.Thus,the dorsal esophagus and the more ventral trachea and lung buds are defined.The larynx,which is formed from the fourth and sixth branchial arches ,maintains communication between the pharynx and trachea.

The lung buds penetrate the coelomic cavity by caudal growth ,resulting in the formation of pleuroperitoneal canals on either side of the foregut.The expanding lung buds eventual come to nearly fill these canals,with the small residual spaces becoming the primitive pleural cavities.

Fetal lung development is divided into five stages; embryonic, pseudoglandular, canalicular, saccular and alveolar (2). Embryonic lung development begins during the third week of gestation as a derivative of the foregut and is marked by the formation of a diverticulum off of the caudal end of the laryngotracheal groove (3). The trachea and the two primary lung buds form from this diverticulum by the fourth week of gestation. At 6 weeks, these lung buds have further developed into defined lobar structures. The pseudoglandular phase of lung development takes place during the 7^{th} to 16^{th} weeks of gestation and involves lung airway differentiation. It is during this period that all bronchial airways develop. From the 16^{th} to the 24^{th} weeks of gestation, fetal lung development enters the canalicular phase of growth. During this period, airspace development occurs, as crude alveolar air sacs begin to take shape. Type 1 pneumocytes ultimately responsible for surfactant production begin to appear.

Gas exchange becomes functionally possible at this stage.Continued maturation of the crude alveolar airspaces takes place during the saccular phase of development that extends from 24 weeks

gestation to term. During this time period there is continued remodeling of the airspace dimensions and a maturation of surfactant synthesis capabilities (4). Mature adult-like alveoli begin to appear shortly after birth (5,6). Extensive alveolar maturation and multiplication then takes place from birth until approximately 8 years of age, with a 10-fold increase in the number of functioning alveoli (7,8,9). Investigators have proposed that alveolar formation may be completed by 2 years of age (10).

Pulmonary vascular development follows the stages of airway and alveolar growth and can be devided into two anatomic units based on associated airway structure. The term acinus describes the functional unit of the lung that includes the respiratory bronchioli, alveolar ducts, and alveoli- all structures that evolve during or after the canalicular phase of lung development. Vascular development in this region proceeds concurrently with alveolar growth and multiplication. The preacinar structures include the trachea, major pronchi, and lobarbronchi up to the terminal bronchioles. Preacinar vascular development is completed by 16 weeks' gestational age(11,12,13).

FACTORS GOVERNING LUNG DEVELOPMENT

It is now recognized that pulmonary development is marked by a series of programmed events regulated by master genes such as the homeobox genes, nuclear transcription factors, hormones, and growth

factors. These processes involve genes regulating epithelial and endothelial interactions as well as temporal and spatial interactions if several hormones and growth factors. Early development transcription factors such as hepatocyte nuclear factor-3β and thyroid transcription factor-I regulate pulmonary development from the foregut mesenchyme. Additional stimuli of pulmonary development involve the transforming growth factor- β pathway, sonic hedgehog pathway, Notch-delta pathway, Wingless-Int pathway ,and cytokine receptor pathways. Subsequent signal transduction control of organogenesis includes the apoptotic pathways, nuclear receptor pathways, and interleukin pathways. Hormones such as the glucocorticoids, thyroid hormone, and retinoic acid have been shown to regulate several of the crucial cellular interactions required for proper pulmonary organogenesis and differentiation(14,15,16,17,18,19).

HISTORY

Congenital cystic adenomatoid malformation was first described as distinct pathologic entity by Chin and Tang in 1949(20).Before then CCAM was grouped under the general diagnosis of congenital cystic lung disease ,along with bronchopulmonary sequestration , congenital lobar emphysema and bronchogenic cyst. First ultrasonic detection of CCAM was done in the year 1975 by Garret WJ et al

INCIDENCE

The true of cystic lesions is unknown because there are no population based studies in literature. Recent studies indicate the incidence of CCAM in the range of 1:35000 to 1:10000. CCAM is the most common chest mass detected in the fetus and accounts for more than 25% of congenital lung lesions

PRENATAL DIAGNOSIS

Prenatal diagnosis provides insight into the in utero evolution of fetal lung lesions such as congenital cystic adenomatoid malformation (CCAM) ,broncho pulmonary malformation and congenital lobar emphysema. Serial sonographic study of fetuses with lung lesions has helped define the natural history of these lesions ,determine the pathophysiologic factors that determine the clinical outcome and formulate management based on prognosis(21,22,23,24,25,26). In prenatal ultrasonogram it is seen as an echogenic lesion.A series of more than 175 prenatally diagnosed cases from the children's hospital of Philadelphia and University of California ,SanFransisco found that the overall prognosis depends on the size of lung mass and the secondary physiologic derangement: a large mass causes mediastinal shift , hypoplasia of normal lung tissue ,polyhydromnios , and cardiovascular compromise leading to fetal hydrops and death(27).

PATHOPHYSIOLOGY OF FETAL LUNG LESIONS

Huge fetal lung lesions have reproducible pathophysiological effects on the developing fetus. Esophageal compression by the thoracic mass causes interference with fetal swallowing of amniotic fluid and results in polyhydromnios. Polyhydromnios is common maternal condition for pre natal ultrasonography .So , a prenatal diagnostic marker exists for large fetal lung tumors. Support for this concept comes from the absence of fluid in the fetal stomach in some of these cases and the alleviation of fetal hydromnios after effective fetal treatment. The hydrops is secondary to venacaval obstruction and cardiac compression from large tumors causing extreme mediastinal shift. Like CCAMs ,a fetal BPS can also cause fetal hydrops,either from mass effect or from tension hydrothorax that is the result of fluid or lymph secretion from BPS(27).CCAM and BPS form important causes of non-immune hydrops fetalis.Hydrops is a harbinger of fetal or neonatal demise and manifests as fetal ascitis , pleural and pericardial effusions, and skin and scalp edema.Although there is some association of both polyhydromnios and hydrops with fetal lung lesions, experience indicates that either can occur independently of the other.

PROGRESSION IN FETAL LIFE

Large fetal lung tumors may regress in size on serial prenatal sonography illustrating that improvement can occur during fetal life

(28,29,30). In particular many non cystic BPSs dramatically decrease in size and may not need treatment after birth. Many CCAMs also decrease in size on serial prenatal ultrasonogram. However, fetal lung lesions that seem to disappear on prenatal ultrasonogram and are not seen in post-natal chest radiograph still need evaluation with CT scan ,which will frequently detect a lesion(31).

CCAM VOLUME RATIO:

Recently, fetal CCAM volume has been determined by the sonographic measurement using the formula for a prolate ellipse (length x height x width x 0.52). A CCAM volume ratio(CVR) is obtained by dividing CCAM volume by head circumference to correct for fetal size. ACVR greater than 1.6 is predictive of increased risk of hydrops, with 80% of these fetuses developing hydrops.The CVR may be useful in selecting fetuses at risk of hydrops and thus needing close ultrasound observation and possible fetal intervention(32). Serial CVR measurements have shown that CCAM growth usually reaches a plateau by 28weeks of gestation. For fetuses at less than 28 weeks gestation , the recommendation is twice weekly ultrasound surveillance if the CVR is greater than 1.6 and initially weekly surveillance for fetuses with smaller CVR values.

CLASSIFICATION

Major distinctive pathological (and radiological) features allow to classify CCAM into following broad categories.Historically congenital cystic adenomatoid malformations have been divided by Stocker into 3 types.

1. STOCKER TYPE I :large cyst (>2cm)

Currently referred to as the macrocystic CCAMs based on gross anatomy and ultrasound findings(34) .These consist of large sometimes multiple or multiloculated cysts.They are the result of terminal bronchiolar proliferation with associated suppression of alveolar development. They are not true cysts and always communicate with proximal airway and distal lung parenchyma(33). Histologically they are lined with respiratory ciliated cuboidal or columnar epithelium.

2. STOCKER TYPE II

Currently referred to as microcystic CCAM.These consist of small uniform multiple or multiloculated cysts.They are not true cysts and are lined by ciliated columnar or cuboidal epithelium.

3. STOCKER TYPE III:(solid lesion)

These are macroscopically and microscopically solid lesions without cysts. Pathologically these are grouped into pulmonary hyperplasia group. Stocker JT suggested a new name ,congenital pulmonary airway malformation in an article in histopathology journal in the year 2002(36).

BLOOD SUPPLY

CCAM receives its blood supply from the pulmonary circulation and is not sequestered from the tracheobronchial tree. However, type II and III lesions can occasionally coexist with extralobar sequestration(hybrid lesions), and in such cases, they may receive systemic arterial supply

HISTOLOGY

CCAM is differentiated from other congenital cystic diseases by 4 characteristics :

(I) Polypoid projections of the mucosa

(II) An increase in smooth muscle and elastic tissue in cyst walls

(III) Absence of bronchial cartilage (unless it is trapped within the lesion)

(IV) Presence of tall columnar mucinous epithelium

(V) Absence of inflammation (35,58)

MANAGEMENT IN ANTENATAL AND POSTNATAL LIFE:

ANTENATAL DIAGNOSIS

Cystic lung lesions are the most commonly identified pulmonary lesions in prenatal ultrasonography which has proved its diagnostic accuracy in 70% of cases(37).On occasion cystic lung lesion in lung base may be confused with a diaphragmatic hernia ;or a pulmonary sequestration can be confused with CCAM if USG dost not identify systemic feeding vessel in Doppler.In these unclear cases ,ultrafast MRI can provide more detailed anatomic assessment to make a diagnosis(38).Serial prenatal USG is needed in the follow up of these prenatally diagnosed lesions.

POSTNATAL DIAGNOSIS

If the lesion is not identified prenatally, then a two-view chest radiograph is the first diagnostic study in a patient with respiratory symptoms; but it can appear normal and, unless a high index of suspicion is maintained, the diagnosis can be missed.(39) If an abnormality is identified on chest radiography, or if the chest radiograph is normal but the patient is symptomatic, then a confirmatory study can be performed next. The choice of study is dependent on the patient's symptoms and likely diagnosis. Confirmatory studies include CT scan (hyperinflation is better seen, systemic arterial supply to sequestrations seen in 60% of the cases), 7 ultrasonography, and contrast esophagography (to demonstrate

communication between the respiratory tract and esophagus or stomach in sequestration for example (40) Occasionally, magnetic resonance imaging and bronchoscopy are necessary (41). Rarely is angiography needed to make the diagnosis or aid in preoperative planning particularly in cases of severe inflammation. Its potential complications outweigh its benefits especially in infants. (42) In such cases, MR angiography may be the study of choice. (43,44)

ROLE OF FETAL INTERVENTION

The finding that fetuses with hydrops are at very high risk for fetal or neonatal demise led to the performance of either fetal surgical resection of the massively enlarged pulmonary lobe (fetal lobectomy) for cystic /solid lesions or thorcoamniotic shunting for lesions for lesions with a dominant cyst(45,46). Lesions with associated hydrops that are diagnosed late in gestation may benefit from resection using an Ex-utero Intrapartum therapy approach(EXIT)(47) .The fetus with a lung mass but without hydrops has an excellent chance for survival with maternal transport ,planned delivery ,and neonatal evaluation and surgery.

A few fetuses may develop fluid collections within the chest cavity and in those situations a Harrison catheter shunt can beused to drain the fluid into the amniotic fluid. Very large cystic masses might pose a danger during birth because of the airway compression. In this situation, a

special surgical type of delivery called the EXIT procedure can be done(48).

In rare extreme cases, where fetus's heart is in danger, fetal surgery can be performed to remove the CCAM. If non-immune hydrops fetalis develop, there is a near universal mortality of the fetus. Fetal surgery can improve the survival.

NEONATAL SURGERY

Neonates with respiratory compromise due to a cystic lung lesions require prompt surgical resection ,usually by lobectomy. In most severe cases, ventilatory support with high frequency ventilation or extracorporeal membrane oxygenation may be required.

MANAGEMENT OF ASYMPTOMATIC PATIENTS

In asymptomatic neonates with a cystic lung lesion ,it is believed that elective resection is warranted because of the risks of infection and occult malignant transformation(71).

AGE OF SURGICAL RESECTION

Postnatally CCAM is confirmed by contrast CT scan .Most experts suggest elective resection of CCAM at one month of age or later.

MORTALITY/MORBIDITY

The prognosis primarily depends on the size of the lesion. In a Canadian series of 48 patients, the incidence of postnatal demise was 10% (10 of 40 patients) with 8 spontaneous and voluntary abortions. Larger lesions have a higher incidence of mediastinal shift, vascular compromise, polyhydramnios, pulmonary hypoplasia, and hydrops, which may lead to intrauterine fetal demise or neonatal death.

The most commonly associated anomalies occur in the type II form. The anomalies affect the renal (cystic disease, agenesis dysgenesis), intestinal (atresias), cardiac, and osseous systems.

Type III CCAM tends to be extensive and therefore tends to have a poor prognosis. The prognosis is also poor with bilateral lung involvement, prematurity, and severe associated malformations.

MALIGNANCIES ARISING FROM CCAM

Children with CCAM have increased risk of developing malignancy.Malignancy mainly consist of pulmonaryblastoma and rhabdomyosarcoma in infants and young children and bronchoalveolar carcinoma in older children and adults(49,50,51,52,53).

DIFFERENTIAL DIAGNOSIS FOR CCAM

The differential diagnosis for fetal lung space occupying lesions include

A) Diaphragmatic hernia,

B) Pulmonary sequestration,

C) Bronchogenic cysts, and

D) Congenital lobar emphysema.

DIAPHRAGMATIC HERNIA

Congenital diaphragmatic hernia (CDH) is a rare congenital anomaly characterized by a defect in diaphragm development with subsequent herniation of abdominal contents into the thorax. The two most prevalent types are herniation anteriorly in the diaphragm (Morgagni hernia) and posterolateral (Bochdalek-type) hernias. The incidence of posterolateral CDH has been reported from between 1 in 2,000 and 1 in 5,000 live births (54). Classically, infants with CDH present in respiratory distress either at birth or within the first few hours of life. Currently, between 40% and 60% of infants with CDH are diagnosed prenatally (55).

The diagnosis of congenital diaphragmatic hernia is suspected when the prenatal ultrasound demonstrates the heart in an abnormal

location and fluid-filled loops of bowel are visualized in the thorax. The diagnosis can be confused with a cystic adenomatoid malformation of the lung, and a fetal magnetic resonance imaging may be helpful to distinguish the two (56) In infants who present at or after delivery, respiratory distress is the main complaint. The infant will often have a scaphoid abdomen. There may be bowel sounds in the chest. Location of the nasogastric tube in the thorax on radiograph or an upper gastrointestinal contrast study will aid in confirming the diagnosis.Treatment for congenital diaphragmatic hernia is surgery.Recent trend is to first stabilize the patient ,and surgical repair at an appropriate time.

BRONCHOPULMONARY SEQUESTRATION

Pulmonary sequestrations make up10-30% of the cystic bronchopulmonary foregut malformations. Bronchopulmonary sequestrtions (BPS) may be intralobar or extralobar sequestration.In both types of BPS there is no communication between the sequestrum and the normal tracheo-branchial tree.On prenatal USG ,a BPS appears as a well defined echodense,homogenous mass.Detection by color flow Doppler of a systemic artery or arteries from the aorta to the fetal lung lesion is pathognomonic of BPS(59).However if this finding is not detected ,an echodense CCAM and a BPS can have similar prenatal sonographic appearance.Ultrafast fetal magnetic resonance imaging can differentiate CCAM and BPS(60). Furthermore there are prenatally detected lung

masses that display clinicopathogenic features of both CCAM and BPS .These are called as hybrid lesions denoting shared embryonic basis for these lung lesions(61,62,63). An intralobar sequestration is most commonly seen in the medial basal or posterior basal segments of the lower lobes, left side more common than the right side. Usual postnatal presentation of intralobar BPS is recurrent pneumonia and even abscess formation with in the BPS. Extralobar BPS is more frequent in males ,is more common on the left side. Extralobar BPS is associated with conditions such as congenital diaphragmatic hernia,vertebral anomalies and congenital heart disease.

CONGENITAL LOBAR EMPHYSEMA

Fundamental mechanism of congenital lobar emphysema(CLE) is that the affected bronchus allows passage of air on inspiration but only limited expulsion of air on expiration leading to lobar overexpansion. Air trapping in CLE lobe may be due to (a) dysplastic bronchial cartilages creating a ball valve effect or a complete bronchial atresia; (64,65) (b) endobronchial obstruction from inspissated mucus or extensive mucosal proliferation and infolding; (66) (3) extrinsic compression of the bronchi from aberrant cardiopulmonary vasculature or enlarged cardiac chambers;(67) and (4) diffuse bronchial abnormalities that may or may not be related to infection(68).The most common site of involvement for CLE is the left upper lobe (40% to 50%), followed by the right middle lobe (30% to 40%) and right upper lobe (20%).Congenital lobar

emphysema can be distinguished prenatally from other cystic lung lesions on ultrasonography by increased echogenicity reflectivity compared with a microcystic CCAM and the absence of systemic arterial blood supply compared with a BPS(69,70).Progressive enlargement of these lesions prior to 28 weeks gestation may be due to fetal lung fluid trapping in the lobe analogous to the air trapping seen postnatally. Late in the gestation CLE may regress in size and the character of the mass ,rendering it indistinguishable from adjacent normal fetal lung. Postnatal assessment is important is because of postnatal air trapping in the emphysematous lobe. At the time of birth the affected lobe may be radioopaque on chest X-ray because of the delayed clearance of fetal lung fluid. CLE is diagnosed at birth in about 25% of cases and by the age 1 month in about 50%.The diagnosis is sporadic after 6 months of age. If the presentation is respiratory distress and pulmonary lobar hyperinflation ,then the main stay of management is resection of emphysematous lobe.

CONGENITAL BRONCHOGENIC LUNG CYSTS

Bronchogenic cysts are typically thick walled,unilocular lesions which are comprised of smooth muscle ,cartilage and mucous glands lined by pseudostratified ciliated columnar epithelium.It is believed that they become separated from the tracheobranchial tree during development,but remain adjacent ,which is they are found clinically. Congenital lung cysts may develop at any time between the third and the 16^{th} weeks of gestation as the lung buds begintheir initial segmental

divisions and subsegmental dichotomous divisions progress.Bronchogenic cysts arise from the trachea ,bronchus and other conducting airways but have usually lost their connection with their parent structure.They are usually simple ,contain mucous however, air fluid levels and infection may be seen if there is continuity with tracheobronchial tree .Majority of bronchogenic cysts are found in the lung parenchyma and mediastinum. If symptomatic most common presentations are wheezing, tachypnoea or dyspnoea ,all related to compression of the adjacent conducting airway with partial obstruction.If there is patent connection with tracheobronchial tree ,the may get infected and the child may present with features of infection. CT or MRI imaging will allow these lesions to be differentiated from other congenital cystic lung lesions.

PATIENTS AND METHODS

It is a combined prospective and retrospective study which included patients with congenital cystic adenomatoid malformation, who attended the pediatric surgery OPD at the Institute of Child Health and Hospital for Children, Madras Medical College, Chennai. The study was done during the three years period, from Jan 1, 2007 to Dec 31, 2009.

Selection Criteria

Inclusion Criteria

All patients had proven congenital cystic adenomatoid malformation radiologically.

Exclusion Criteria

- All cases of bronchopulmonary sequestration
- All cases of congenital lobar emphysema
- All mediastinal cystic lesions like bronchogenic cyst, neurenteric cyst, cystic teratoma and pericardial cyst

The patients were subjected to detailed clinical examination and relevant investigations were performed, namely, chest x-ray and CT scan chest .

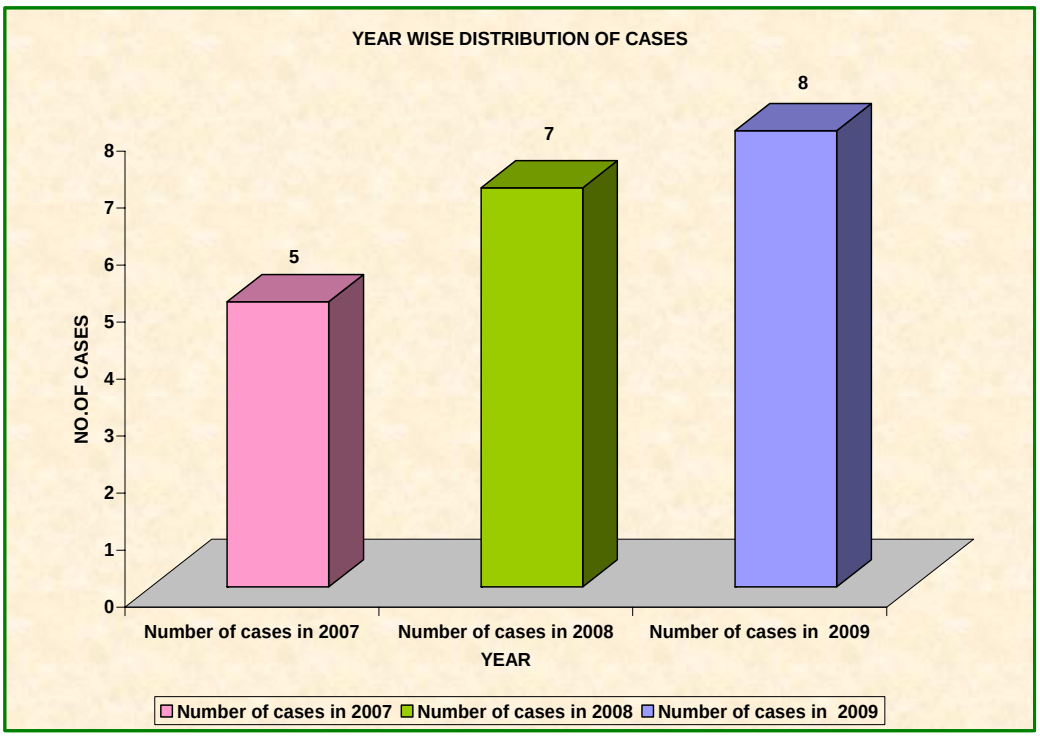
The treatment modalities were studied and patients were followed up to assess the effectiveness after 6 months of surgery with relevant investigations and extended to the available period. The results were tabulated and analyzed

OBSERVATIONS

This study of congenital cystic adenomatoid malformation presented to Pediatric Surgery Department , ICH&HC, MMC, Chennai was undertaken between January 2007 to December 2009. The following facts were obtained:

During the study period of 3 years from January 2007 to December 2009, twenty patients fulfilled the criteria. Out of these 20 patients ,five patients presented in 2007 ,seven patients presented in 2008 and eight patients presented in 2009.

Number of cases in 2007	5
Number of cases in 2008	7
Number of cases in 2009	8
Total	20


YEAR WISE DISTRIBUTION OF CASES
NO.OF CASES
0
1
2
3
4
5
6
7
8
5
7
8
Number of cases in 2007
Number of cases in 2008
Number of cases in 2009
YEAR
Number of cases in 2007
Number of cases in 2008
Number of cases in 2009

ANTENATALLY DETECTED CASES

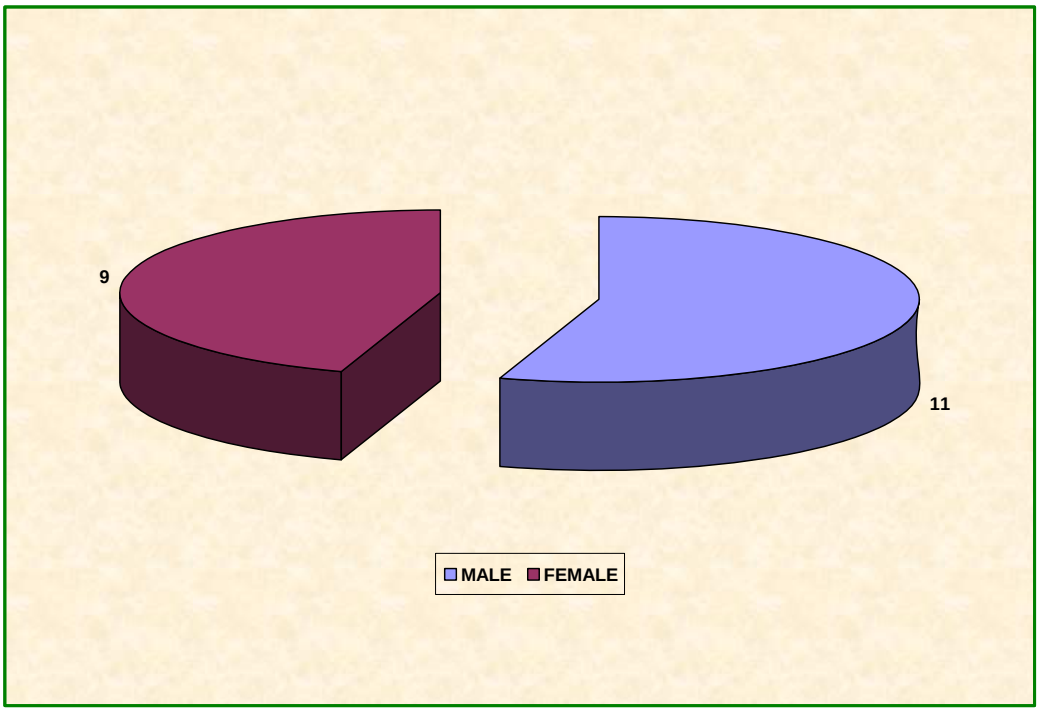
Out of these twenty patients, seven were ante-natally diagnosed. Among these cases five patients presented in 2009, one each in 2007 and 2008.This may be due to increase in the efficiency and familiarity of the sonologists in dealing with fetal lung lesions.Majority (five) of antenatally detected patients were asymptomatic at the time of presentation.

Remaining two patients were admitted with complaints of having respiratory distress since birth .One patient was admitted soon after birth ,and was put on mechanical ventilation.Child was on ventilator for 23 days and expired.The other one presented on 34th day of life and was evaluated and successfully operated.

SEX DISTRIBUTION

In this study, 11 patients were male and the remaining 9 were female.There is no sex predilection for patients with CCAM.

MALE	11
FEMALE	9

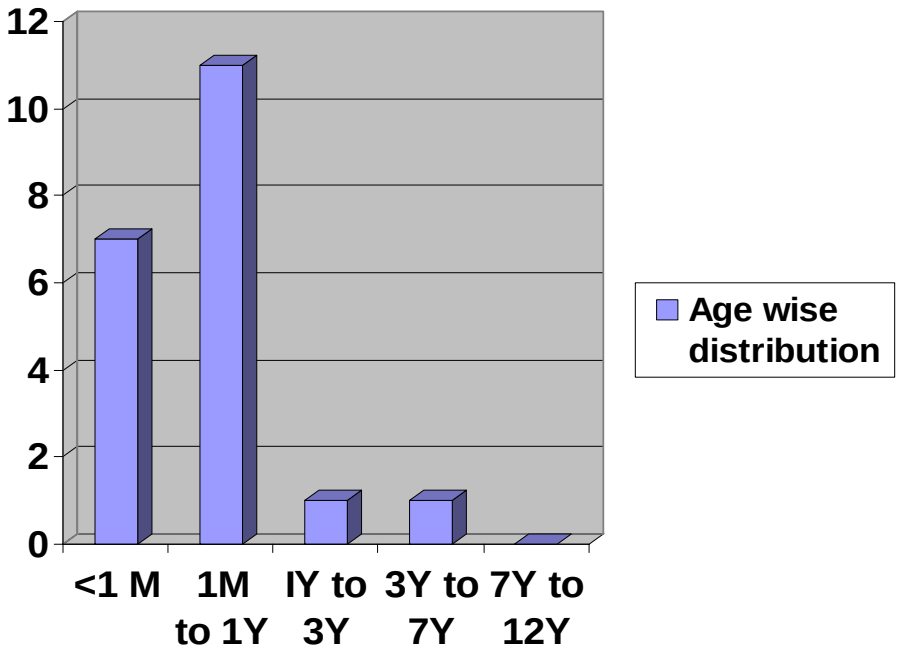


AGE OF PRESENTATION

Among our study group of twenty patients , 7 patients presented in the neonatal period ,11 presented between 1 month and 1 year,2 patients presented between 1 to 5 years .No patient was seen above 5 years of age.This means that 90% of CCAM patients in our study are below one year of age .High degree of suspicion is needed to detect CCAM in infants if they are not antenatally diagnosed.

UPTO 1 MONTH	7	35%
1 MONTH TO 1 YEAR	11	55%
1 TO 3 YEARS	1	5%
3 TO 7 YEARS	1	5%
7 TO 12 YEARS	0	0%

AGE WISE DISTRIBUTION OF CCAM PATIENTS

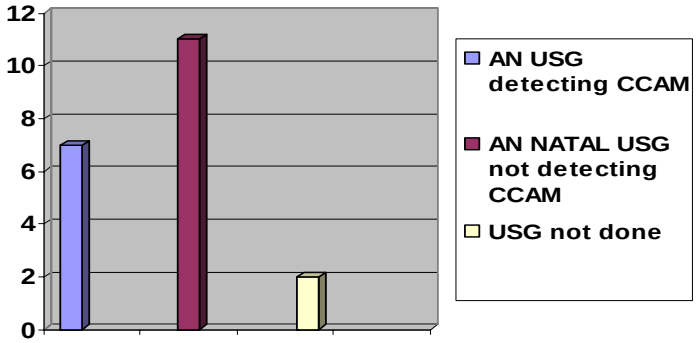


EFFECTIVENESS OF ANTENATAL USG

In the patients who were enrolled for study 18 underwent atleast one antenatal USG.2 patients didn't have any antenatal USG. Among this group of 18,only 7 were suspected to have cystic lesion in lung.This means that only 39% patients were detected antenatally. Internationally, screening antenatal USG detects about 70% of CCAMs.Of the 18 patients who went antenatal USG ,only one patient had polyhydromnios.But this infant was not detected to have CCAM antenatally.

AN USG done patients	18 (90%)
AN USG not done	02 (10%)

AN natal USG detecting CCAM	7 (39%)
AN USG not detecting CCAM	11 (61%)
(Two patients didn't have AN USG)	



MODE OF PRESENTATION

PRESENTING COMPLAINTS	NUMBER OF CASES
Asymptomatic	5
Respiratory distress since birth	3
Fever with cough	6
Dyspnoea with cough	4
Cough with wheeze	2

CCAM presents in various forms. In our study, 5 asymptomatic patients attended surgical OPD.All these five were antenatally detected.

Three patients presented with complaints of respiratory distress since birth.Among these three ,two patients presented on the 1st day of life.One patient with respiratory distress since birth presented on 34th day of life.

Six of our patients presented with fever and cough .Among this group, three were initially treated outside as bronchopneumonia ,Two patients as empyema and one as loculated pyopneumothorax. Four CCAM patients presented with difficulty in breathing and two presented with wheeze.

Patients with empyema and loculated pyopneumothorax were initially treated with intercostal chest tube drainage .Only later they were found to have CCAM.

SENSITIVITY OF CHEST X-RAY

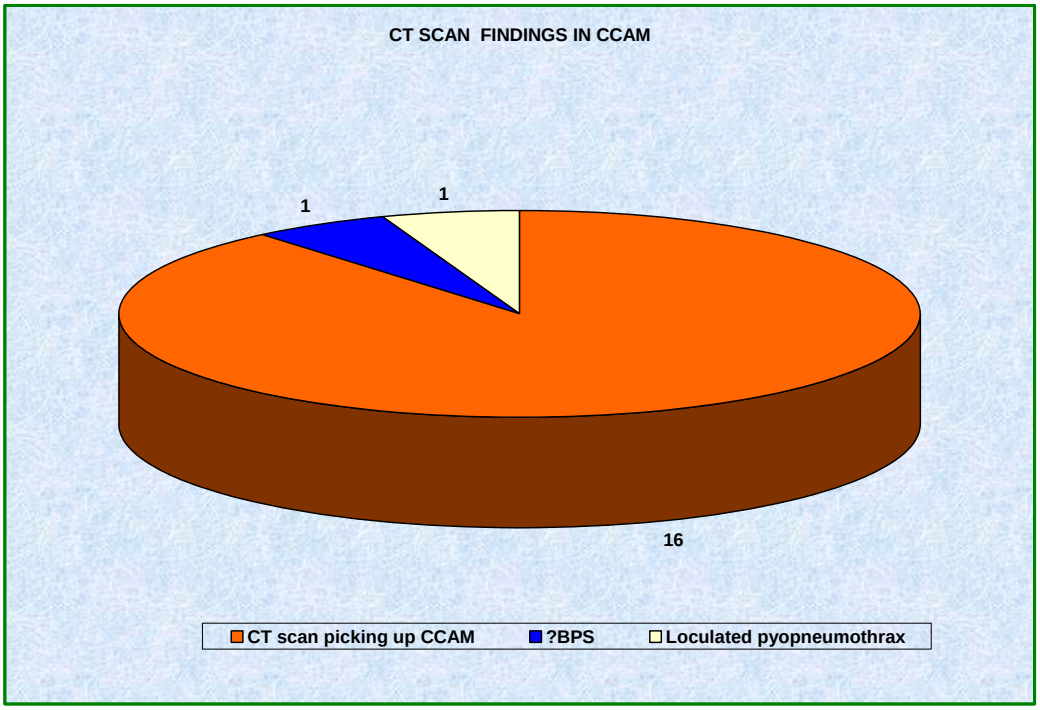
X-RAY diagnosis	
CCAM	10
Bronchopneumonia	5
Lung cyst	2
Empyema	2
Pyopneumothorax	1

All of our patients had chest radiography at the time of admission. Radiologists were able to suggest the diagnosis of CCAM in only 10 patients. For two patients they gave the diagnosis of lung cyst and for five patients they gave the diagnosis of bronchopneumonia. Two patients were diagnosed to have empyema and one was diagnosed to have loculated pyopneumothorax.

ROLE OF CT SCAN CHEST

CT scan was done in 18 of our CCAM patients. Among these children 16 were diagnosed to have CCAM ,one was found to have ?BPS and one was diagnosed as loculated pyopneumothorax.This shows postnatal CT scan has a higher diagnostic value for detecting lung lesions.

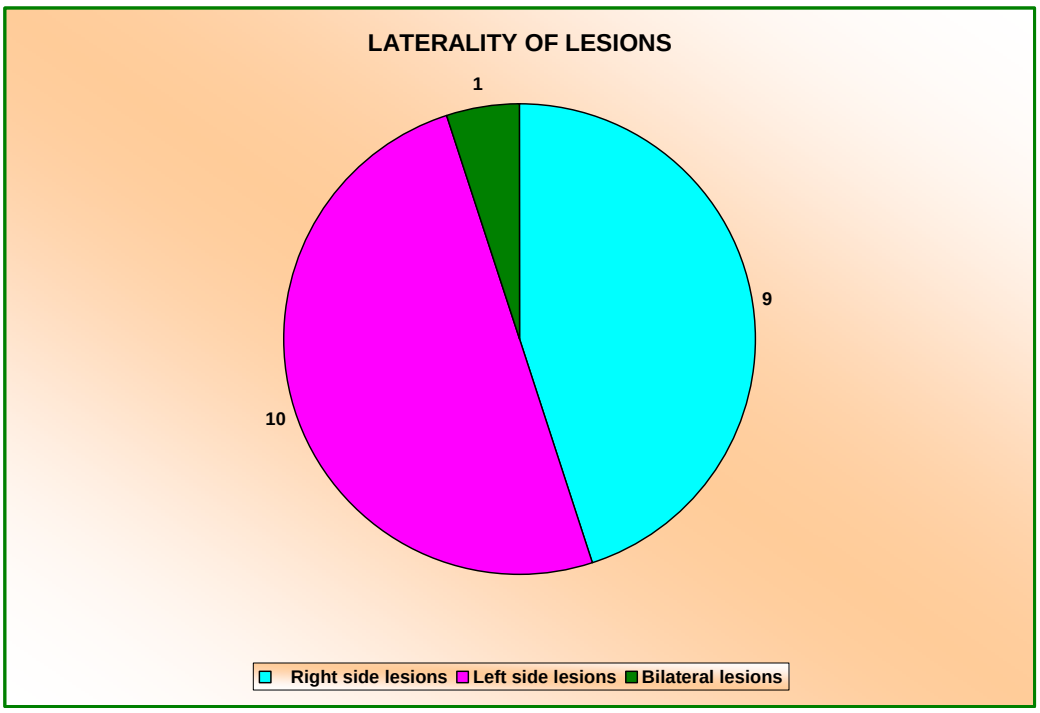
CT scan picking up CCAM	16
?BPS	1
Loculated pyopneumothrax	1



LATERALITY OF LESIONS

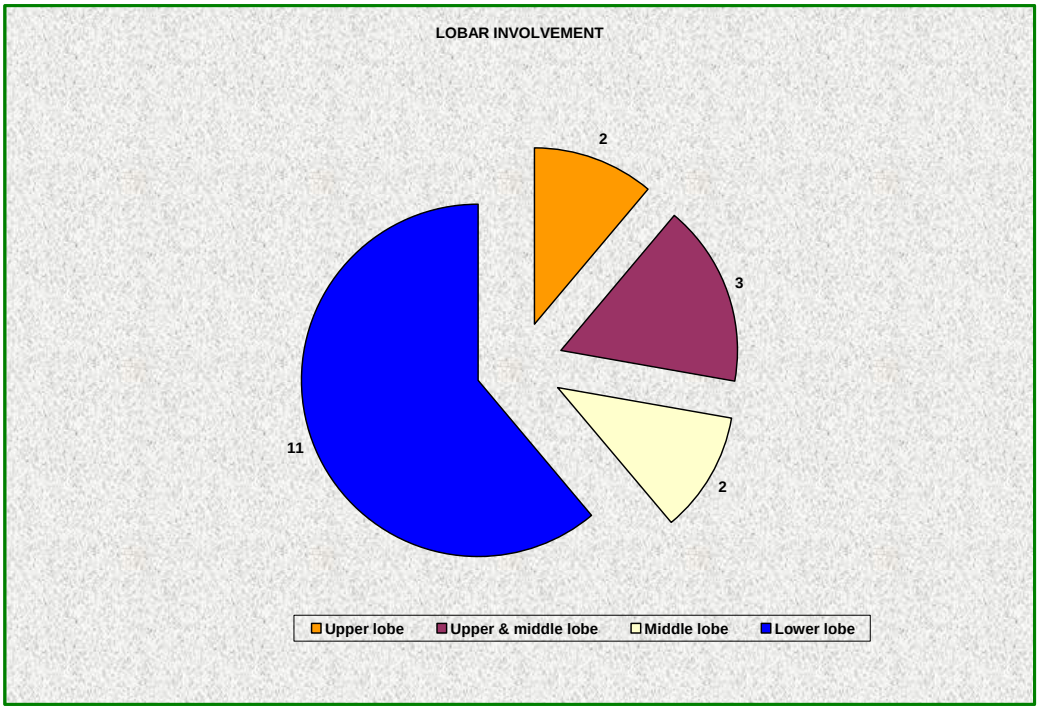
Right side lesions	9 (45%)
Left side lesions	10 (50%)
Bilateral lesions	1 (5%)

In our study group 9 patients had right sided lesions ,10 patients had left sided lesions and 1 had bilateral congenital cystic adenomatoid malformation.



LOBE INVOLVED

Upper lobe	2 (11%)
Upper & middle lobe	3 (17%)
Middle lobe	2 (11%)
Lower lobe	11 (61%)



Lower lobe is the most affected lobe .Lower lobe was affected in 11 patients, upper lobe 2 patients, middle lobe 2 patients and 3 patients had both upper and middle lobe involvement. An article in seminars in paediatric surgery(2008) 17 ,2-8 ;by Christina M.Shanthi also endorses the fact that lower lobe is most commonly affected lobe and there is no left to right side difference in the presentation of CCAM.

Lower lobectomy was done in 10 patients(right-3;left-7) , upper lobectomy was done in 5 patients(right -4 ,left 1) and middle lobectomy was done in 5 patients. Among the above patients combined upper and middle lobectomy was done in 3 patients.One patient who presented with empyema also underwent decortication.

POST OPERATIVE PERIOD

Postoperative period was uneventful in 8 patients. 4 patients had wound infection, 2 patients needed post operative ventilatory support. Antenatal detected patients were operated after one month of age.

Uneventful	11 patients
Wound Infection	4 patients
Post operative Ventilatory support	2 patients

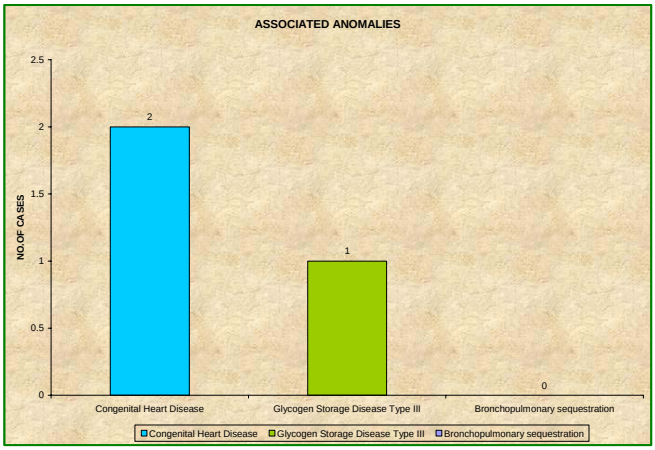
DURATION OF HOSPITAL STAY

Average duration of hospital stay for operated patients was 14.4 days.But the average hospital stay duration of antenatal detected patients was significantly less .It is about 10 days for antenatal detected patients.So it is clear that infected CCAM patients had longer hospital stay.

ASSOCIATED ANOMALIES

In our study 2 patients had congenital heart disease and one patient had glycogen storage disease type III. In our study there was no association with bronchopulmonary sequestration.

Congenital Heart Disease	2
Glycogen Storage Disease Type III	1
Bronchopulmonary sequestration	0



PATHOLOGY REPORT OF LESIONS

Stocker type I	16 cases (94%)
Stocker type III	1 case (6%)

In our series 94% of cases belong to Stocker type I lesion and 6% of cases belong to Stocker type III lesion.

FOLLOW UP

In our series there is no mortality among operated patients Among the 17 patients who we have operated, 15 patients turned up for follow up. Out of this, two patients were treated for respiratory tract infection. Their chest X-ray are normal. Pulmonary function tests are planned for the future.

CONCLUSION

- Most of CCAM patients present early in life (< 1 year)
- In our setup the ability of antenatal USG to detect lung lesions is only 39%
- Antenatally detected patients , report to the hospital earlier than the other patients.
- Presenting symptoms vary widely from mild respiratory tract infection to empyema.
- CT scan chest is the investigation of choice.
- Antenatally detected patients are usually asymptomatic
- Patients with pulmonary infection have a longer hospital stay and have higher incidence of postoperative complications.
- Majority of our patients (94%) belonged to Stocker type I group.
- Our patients are free of respiratory problems in their followup.

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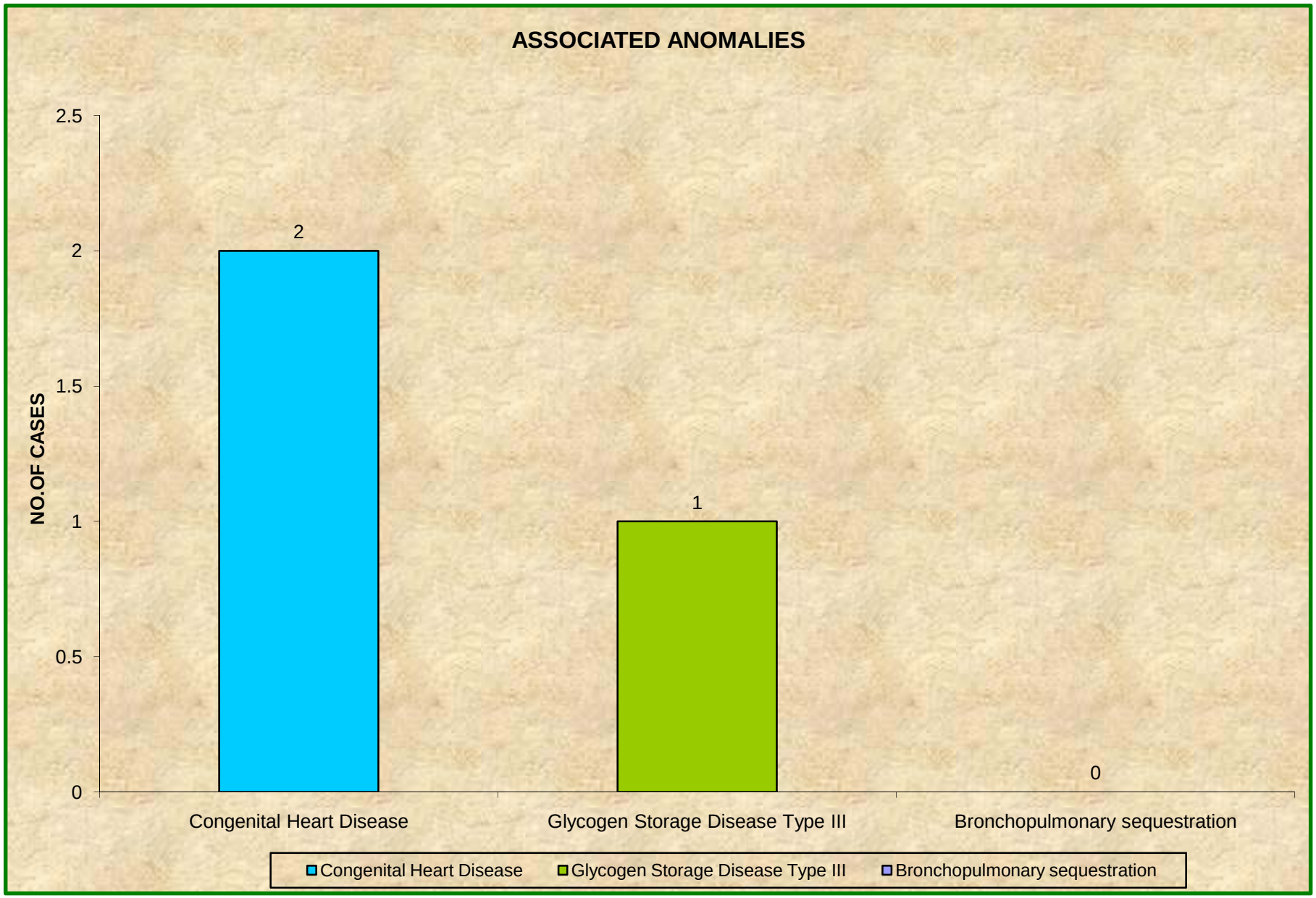
MASTER CHART

Name	IP No	Sex	age of presentation	AN USG	AN CCAM Diag	Presenting Complaints	CXR Diagnosis	CT Scan	Lobe Affected	Side	Treatment	Post OP period	Hospital Stay
ammu	616878	F	6 months	done	not done	bronchopneumonia	Rt lung cyst	CCAM	lower lobe	right	Rt L.Lobectomy	uneventful	12 days
b/o lakshmi	670204	M	14 days	done	not done	dyspnoea-4days;wheeze	s/o CCAM	CCAM	lower lobe	left	Lt.L.lobectomy	change on V POD	22 days
b/o malathi	667307	M	45 days	done	done	asymptomatic	?CCAM ?BPS	CCAM ?BPS	lower lobe	left	Lt.L.lobectomy	uneventful	8 days
b/o mumtaz	670909	F	45 days	done	done	asymptomatic	?CCAM	CCAM	lower lobe	right	Rt L.Lobectomy	uneventful	8 days
ebenezer	657668	M	5 months	done	not done	dyspnoea-4d,fever-2d	Rt lung cyst	CCAM	middle lobe	right	Rt M.Lobectomy	uneventful	21 days
pavithra	641640	F	4 1/2 years	done	not done	recurrent RTI;fever-2 wk	Lt pleural effusion	CCAM	lower lobe	left	Lt.L.lobectomy	wound infection	12 days
savitha	646679	F	11 months	done	not done	recurrent RTI;	bronchopneumonia	CCAM	upper lobe	left	Lt.U.lobectomy	uneventful	10 days
srinivasan	607752	M	15 months	not done	not done	fever-20d;cough&wheeze-3d	bronchopneumonia	CCAM	lower lobe	left	Lt.L.lobectomy	ICD change on III POD	11 days
surya	602012	M	3 months	done	not done	cough-2M;dyspnoea-1 M	bronchopneumonia	CCAM	U&M lobes	right	Rt.U&M.lobectomy	seizures on II POD - dyselectrolytemia	20 days
rasaiya	604871	M	11 months	done	not done	fever-15d;cough-15d	Rt empyema	?CCAM	lower lobe	right	Rt.L.Lobectomy;decortication	wound infection	28 days
raja	595229	M	4 months	done	not done	fever-10d;dyspnoea-7d	Lt pyopneumothrax	loculated pyopneumothorax	lower lobe	left	Lt.L.Lobectomy	wound infection	18 days
nethra	592451	F	34 days	done	done	Resp.distress since birth	CCAM	CCAM	U&M lobes	right	Rt.U&M.lobectomy	iCD change on IV POD	15 days
b/o vanitha	586883	F	8 days	done	done	asymptomatic	CCAM			left	unwilling for surgery		
b/o porkodi	577108	F	28 days	done	done	asymptomatic	B/L CCAM	CCAM	lower lobe	right&left	unwilling for surgery		
rahul	540563	M	28 days	done	not done	Resp.distress since 5th day	bronchopneumonia	CCAM	lower lobe	left	Lt.L.lobectomy with lingula	vent.support-2d	10 days
b/o sathyavathi	527560	M	1st day	done	done	asymptomatic	CCAM	CCAM	middle lobe	right	Rt.M.lobectomy	uneventful	9 days
anantheeswar	586730	M	7 months	done	not done	resp.distress -1 month	CCAM	CCAM	U&M lobes	right	Rt.U&M.lobectomy	uneventful	13 days
swedha	532438	F	8 months	not done	not done	fever&cough-15 days	bronchopneumonia	CCAM	upper lobe	right	Rt.U.lobectomy	wound infection	17 days
b/o harini	658003	M	1 st day	done	done	Resp.distress since birth	CCAM	not done		left	Mech.ventilation for 23 days		
b/o manjula	653115	F	1 st day	done	not done	Resp.distress since birth	CCAM	CCAM	lower lobe	left	Lt.L.lobectomy in 2nd month	uneventful	10 days

Asso Findings
GSD type III
unwilling for surgery
unwilling for surgery
cardiac-VSD
expired ; CHD


ASSOCIATED ANOMALIES
NO.OF CASES
2.5
2
1.5
1
0.5
0
2
1
0
Congenital Heart Disease
Glycogen Storage Disease Type III
Bronchopulmonary sequestration
Congenital Heart Disease
Glycogen Storage Disease Type III
Bronchopulmonary sequestration

PROFORMA

Personal Details -

Name **Age** **Sex**

IP No.

Address

Date

Ante-natal Scan –

□ Done

Report -

□ Not Done -

Clinical Symptoms

□ Asymptomatic □ respiratory distress

□ recurrent LRI □ failure to thrive

□ Empyema □ Others

□ Others

Laterality

□ U/L (Rt/Lt)

□ B/L-

Investigations

BU/SC/SE –

CBC

Blood grouping and typing

Chest X-ray

USG Chest

CT chest

Treatment

□ Surgery -

Surgical Details

Age at surgery

Type

Approach

HPE Report

*Polypoid projections of the mucosa

*An increase in smooth muscle and elastic tissue in cyst wall

*An absence of cartilage

*Presence of mucus secreting cells

*To note the presence of hybrid lesions

Post-op Evaluation

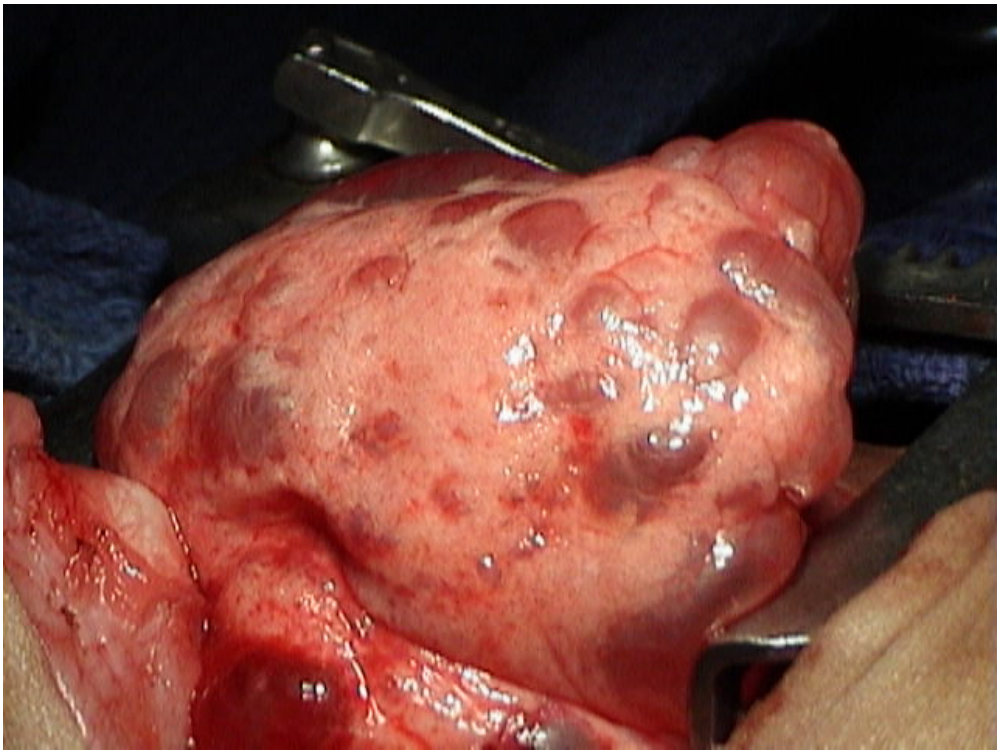
Clinical Symptoms

□ Asymptomatic □ Failure to thrive

□ Recurrent respiratory tract infections

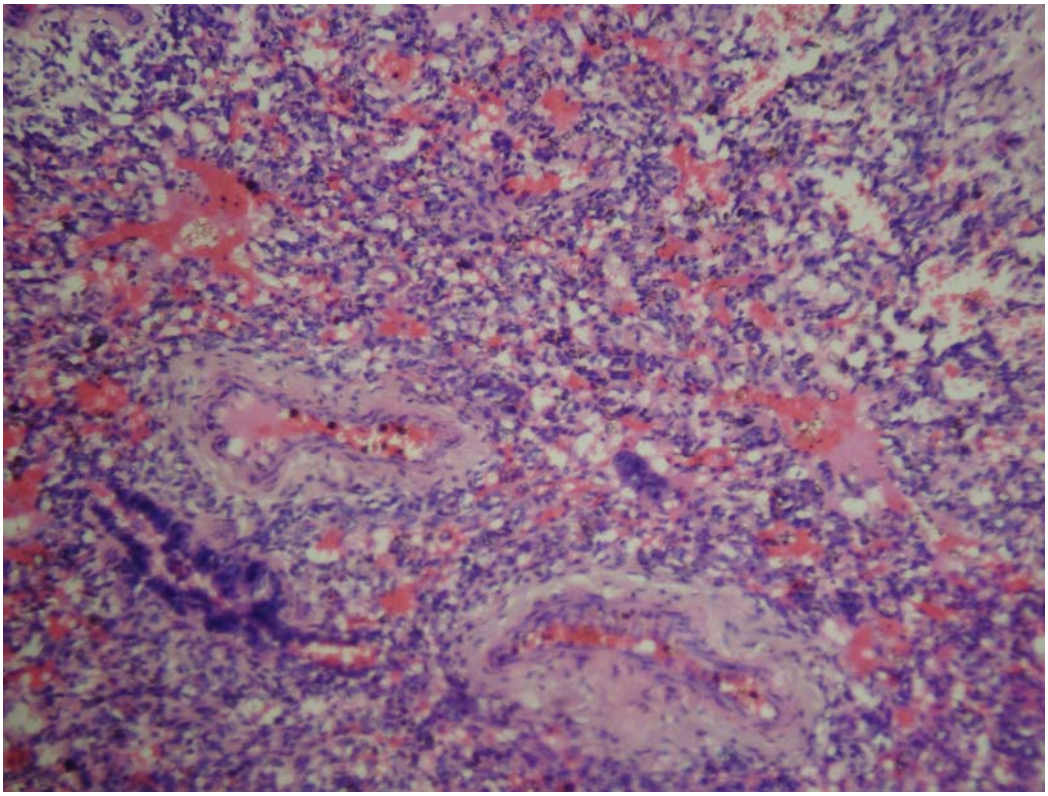
□ Others

CONGENITAL CYSTIC ADENOMATOID MALFORMATION PER OPERATIVE PHOTOGRAPH:

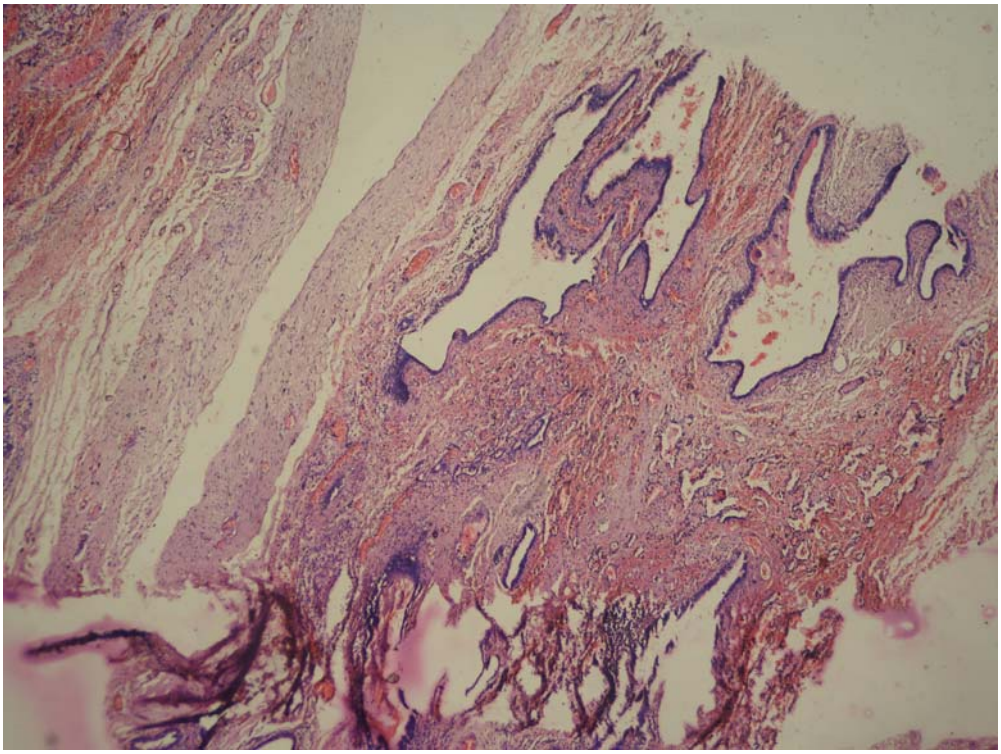


H &E STAINING OF CCAM:

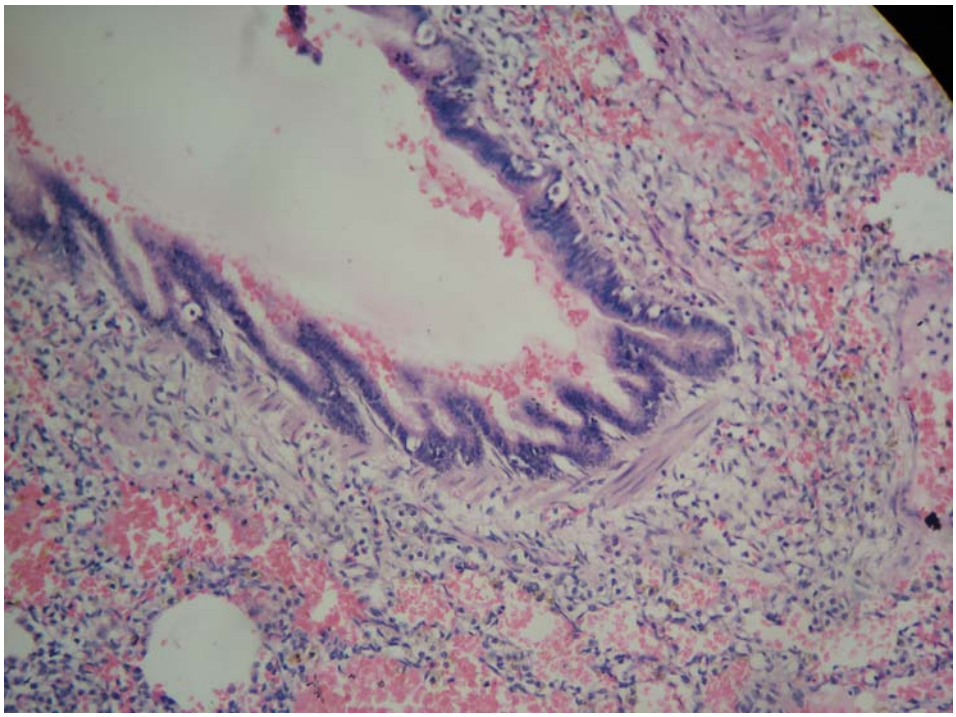
MICRO CYSTIC TYPE OF CCAM



MACRO CYSTIC TYPE OF CCAM



H & E STAIN SHOWING ABUNDANCE OF ELASTIN AND ABSENCE OF CARTILATE



ANTENATAL ULTRASONOGRAPHY OF CCAM:

CHEST RADIOGRAPH OF CCAM PATIENT:

CT SCAN PICTURE OF CCAM PATIENT:

CCAM PRESENTING AS EMPYEMA:

Barium study in CCAM misdiagnosed as CDH in X-ray chest ;